

Asymptomatic Pink Plaque on the Scapula

Cuong V. Nguyen, MD; Daniel D. Miller, MD; Ronda S. Farah, MD



A 36-year-old man presented with a pink plaque on the right side of the scapula of 1 year's duration. The plaque had not grown and was completely asymptomatic. Physical examination revealed a violaceous, pink, 2-cm nodule with overlying telangiectasia. No other concerning lesions were identified on total-body skin examination. A punch biopsy was obtained.

WHAT'S THE DIAGNOSIS?

- a. basal cell carcinoma
- b. cutaneous lymphoid hyperplasia
- c. primary cutaneous follicle center lymphoma
- d. primary cutaneous marginal zone lymphoma
- e. pseudolymphomatous arthropod bite reaction

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From the Department of Dermatology, University of Minnesota, Minneapolis.
The authors report no conflict of interest.
Correspondence: Cuong V. Nguyen, MD, 516 Delaware St SE, Mail Code 98, Phillips-Wangensteen Bldg, Ste 4-240, Minneapolis, MN 55455 (n.cuong87@gmail.com).

THE DIAGNOSIS:

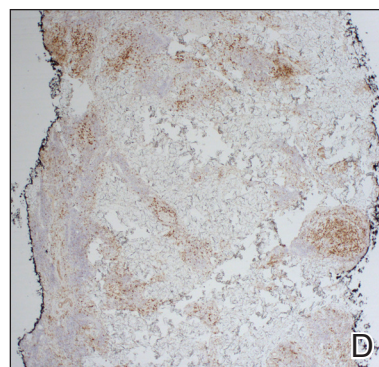
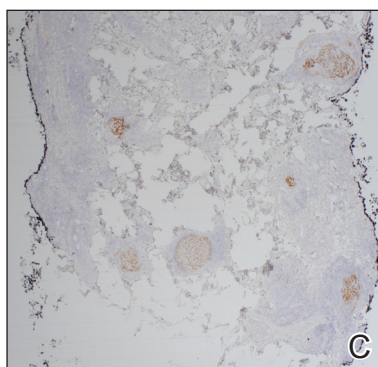
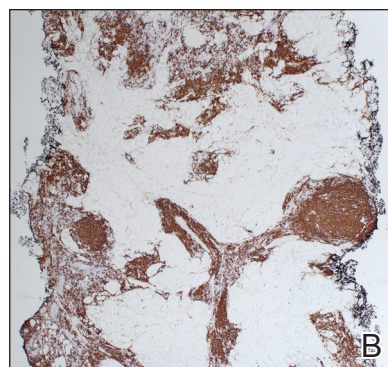
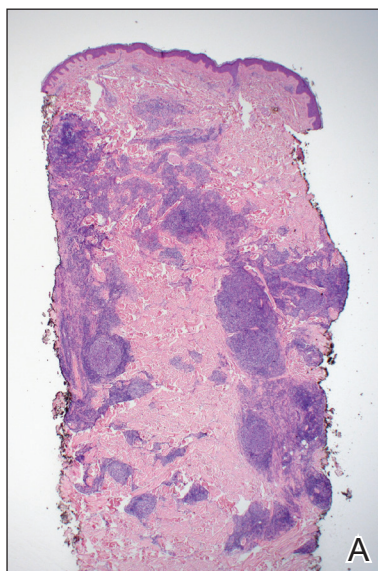
Primary Cutaneous Follicle Center Lymphoma

Immunohistochemistry revealed a nodular infiltrate consisting of small to large atypical lymphocytes forming an irregular germinal center with notably thinned mantle zones and lack of polarization (Figure, A). Atypical cells stained positively with Bcl-6, and CD20 was diffusely positive (Figure, B–D). Bcl-2 and CD3 colocalized to the reactive T-cell infiltrate, and CD10 was largely negative. Further workup with bone marrow biopsy and full-body positron emission tomography–computed tomography was unremarkable. Given these findings, a diagnosis of primary cutaneous follicle center lymphoma (FCL) was made. At 1 month following radiation therapy, complete clinical clearance of the lymphoma was achieved.

Follicle center lymphoma, also known as cutaneous follicular lymphoma, is the most common subtype of primary cutaneous B-cell lymphomas, representing approximately 57% of cases.¹ Follicle center lymphoma typically

affects older, non-Hispanic white adults with a median age of onset of 60 years. It has a predilection for the head, neck, and trunk.² Lesions present as solitary erythematous to violaceous papules, plaques, or nodules, but they can more rarely be multifocal.³ Clinical diagnosis of FCL can be difficult, with papular lesions resembling acne, rosacea, folliculitis, or arthropod assault.^{4,5} As such, diagnosis of FCL typically relies on histopathologic analysis.

Histologically, FCL can present in several different patterns including follicular, nodular, diffuse, or a pleomorphic mix of these.^{2,6} The cells are comprised of germinal center B cells, staining positively for Bcl-6, CD20, and CD79a.⁷ Tumor cells do not exhibit the t(14;18) translocation seen in nodal follicular lymphomas.^{2,8} Unlike marginal zone lymphoma, FCL stains negatively for Bcl-2 and multiple myeloma 1/interferon regulatory factor 4 (MUM1/IRF-4).^{2,9} Forkhead box P1 (FOXP1) also is



Primary cutaneous follicle center lymphoma histopathology revealed nodular and diffuse lymphocytic infiltrate with germinal center formation (A)(H&E, original magnification $\times 20$). CD20 immunostain labeled the majority of the infiltrate (B) (original magnification $\times 40$). CD21 stained follicular dendritic cells and highlighted germinal centers (C)(original magnification $\times 40$). Bcl-6 stained many extrafollicular cells in clusters. Staining was extensive outside the zones of CD21 staining, especially in the top half (D)(original magnification $\times 40$).

usually negative, but its presence can indicate a poorer prognosis.² It is important to distinguish primary cutaneous B-cell lymphomas from systemic B-cell lymphoma with secondary cutaneous involvement, as they have a different clinical prognosis and management course. Further workup includes bone marrow biopsy, serum analysis for clonal involvement, and positron emission tomography-computed tomography imaging. Follicle center lymphoma generally has an indolent disease course with a favorable 5-year survival rate of approximately 95%.^{6,8}

Untreated lesions may enlarge slowly or even spontaneously involute.¹⁰ The histologic growth pattern and number of lesions do not affect prognosis, but presence on the legs has a 5-year survival rate of 41%.² Extracutaneous dissemination can occur in 5% to 10% of cases.² Given the slow progression of FCL, conservative management with observation is an option. However, curative treatment can be reasonably attempted for solitary lesions by excision or radiation. Treatment of FCL often can be complicated by its predilection for the head and neck. Other treatment modalities include topical steroids, imiquimod, nitrogen mustard, and bexarotene.¹⁰ More generalized involvement may require systemic therapy with rituximab or chemotherapy. Recurrence after therapy is common, reported in 46.5% of patients, but does not affect prognosis.²

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